Cardiac Anomalies

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A CLINICOPATHOLOGIC CORRELATION

by

VINCENT MORAGUES M D

Associate Professor of Pathology Creighton University School of Medicine Associate Pathologist, Creighton Memorial St Joseph's Hospital Omaha Nebraska Formerly Associate Professor of Pathology St Louis University School of Medicine St Louis

and

CHESTER P LYNXWILER, M D

Assistant Professor of Pediatrics St Louis University School of Medicine St Louis Director Pediatric Heart Clinic Firmin Desloge Hospital St Louis



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Preface

Congenital cardiac malformations have been in the medical limelight for the past few years. The reason for this special interest has been the great success in the surgical treatment of some of these anomalies. Moreover, increasing numbers of cardiac defects are becoming operable as new surgical techniques are being developed.

In order to give the individual patient the best that surgery can offer, it is necessary that an accurate diagnosis be made in each instance. To accomplish this, however, physical findings must be correlated with the radiologic, physiologic and electrocardiographic findings.

Since the problem is fundamentally one of abnormal morphology of the heart, we have based the classification and the entire correlation within each entity on the features shown by the actual cardiac specimens. We have used exclusively material from cases verified either at surgery or at post mortem examination.

Each anomaly has been covered with brief statements regarding the pathology, course of the circulation, clinical features, laboratory findings, electrocardiography, radiological studies, prognosis, as well as the medical and surgical treatment. Thus we hope this book will be of value not only to the medical student, but to the pediatrician and general practitioner as well.

A unique feature of the book is the presentation of microphotographs of the lungs and great vessels. The changes produced in these structures by certain cardiac anomalies are frequently severe enough to influence the prognosis and the surgical management.

We wish to express our thanks to the physicians of St. Louis and the State of Missouri for sending us many interesting cases as well as many intriguing autopsy specimens. Our appreciation is also extended to the Department of Radiology of the St. Mary's Group of Hospitals of St. Louis University for its excellent cooperation. The assistance of Dr. N. Staples, T. D. Brewer, A. Freitag and Mrs. James Donahoe in preparing the illustrations is sincerely appreciated.

VINCENT MORAGUES
CHESTER P. LYNXWILER

Acknowledgements

We wish to thank the publishers of the below listed journals for allowing us to use figures from the following articles

Figures 22, 23, 24, 25 from V Moragues, "Persistent Common Atrioventricular Ostium," American Heart Journal, Vol 25, No 1, pages 123–127, January, 1943

Figures 34, 35, 36, 37, 38, from Ebel, Lynxwiler and Moragues, "Tricuspid Atresia with Rudimentary Right Ventricle," Journal of Pediatrics, Vol 39, No 2, pages 211–215, August, 1951

Figures 41, 42 from Inkley, Moragues and Lynxwiler, "Tricuspid Atresia," St Louis University Bulletin Vol 2, No 6, June, 1950

Figures 65, 66, 67, 68, 69, 70, 71, 72, 73 from V Moragues, "Persistent Truncus Arteriosus," American Journal of Clinical Pathology, Vol 20, No 9, September, 1950

Figures 130, 131 from Lynxwiler, Smith and Babich, "Coarctation of the Aorta," Archives of Pediatrics, Vol 68 No 5, pages 203–207, May, 1951

Figures 132, 133, 134 from Lynxwiler, Smith, Lucido and Inkley, "Coarctation of the Aorta," Journal of the Missouri State Medical Association, pages 177–180 March, 1951

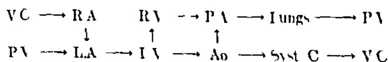
Figures 139, 140, 141 from Moragues Moore and Rossen, "Coarctation of Aorta," American Heart Journal, Vol 24, No 6, pages 828–834, December, 1942

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Introduction

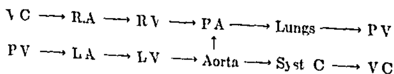
During fetal life the foramen ovale and the ductus arteriosus provide shunting channels through which the pulmonary circulation is bypassed to a great extent. In reality the fetal circulation can be considered as consisting of a single circuit. It is no wonder therefore that even the most formidable malformations of the heart are seldom a cause of intrauterine death. At birth however with the ligation of the umbilical cord and the expansion of the lungs the circulation changes from a single circuit to a double circuit in parallel (systemic and pulmonary). This change entails enormous hemodynamic readjustments which are not possible in the more severely deformed hearts. *This is the reason why the greatest mortality from congenital heart disease occurs in the neonatal period.*

In general there are three mechanisms of adaptation by which the circulation adjusts itself to the altered morphology present in the heart. The *first* of these mechanisms is *complete pooling* of the arterial and venous blood. This mechanism operates in such malformations as cor triloculare, cor biloculare, tricuspid atresia, pulmonary atresia and atresia of the aortic or mitral valve. Complete pooling as a mechanism of adaptation is understood when one considers the course of the circulation in tricuspid valve atresia.



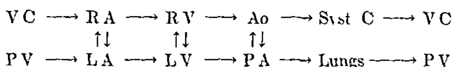
As the tricuspid valve is atretic all the blood entering the right auricle must pass to the left auricle either through an auricular septal defect or a patent foramen ovale. Therefore there is complete pooling of arterial and venous blood in the left side of the heart.

The second mechanism of adaptation is that of a *unidirectional* shunt which occurs in cases of persistent ductus arteriosus, isolated septal defects, and in cases in which the pulmonary artery or aorta "over ride" a ventricular septal defect. In such instances the unidirectional shunts are well tolerated as the shunted blood returns to the side of the circulation from which it started. Unidirectional shunts are easily understood when one considers the course of the circulation in a case of patent ductus arteriosus.



Since the pressure in the aorta exceeds the pressure in the pulmonary artery, the flow through the patent ductus is from the systemic to the pulmonary circulation The blood shunted through the ductus returns to the systemic (left) side of the heart through the pulmonary veins, hence, hemodynamic *equilibrium is maintained*

Multidirectional shunts represent the *third* mechanism by which circulatory adaptation occurs These multiple shunts are necessary in cases of complete *transposition of the arterial* trunks, since in these conditions the shunted blood fails to return to its original side of the circulation To illustrate this mechanism, let us consider the course of the circulation in a case of complete transposition of the arterial trunks with associated septal defects and a patent ductus arteriosus



Since in this *condition* the two *circulations* exist as *entirely* separate circuits, life can be maintained only by the shunting of sufficient oxygenated blood into the systemic circulation However since the shunted blood does not return to the side of the circulation from which it originated, reverse shunts are necessary to maintain blood volume equilibrium in the two circulations It is well known that patients with complete transposition of the arterial trunks live longer when more than one shunt is present

In our opinion, the above mechanisms of circulatory adapta*tion are of such importance in the understanding of the hemo*dynamics in each individual case that they will be referred to again in the various chapters of this book

Most malformations of the heart result in alterations in the pulmonary circulation which interfere with the proper oxygenation of the blood Some of the conditions result in a decreased flow and a decreased pressure in the pulmonary circuit with a consequent oxygen unsaturation of the arterial *blood from* birth (congenital cyanosis) Examples Tetralogy of Fallot, Pulmonary valvular atresia Tricuspid atresia etc Other malformations result in an increased flow and an increased pressure in the pulmonary circulation with subsequent development of pulmonary arteriosclerosis (delayed cyanosis) Examples occasional Patent ductus arteriosus Eisenmenger's complex etc

To give objective evidence of the changes in the pulmonary circulation in these conditions a number of lung sections and *sections* of the aorta and the pulmonary artery have been included in this atlas

1 Malpositions of the Heart

Pathology

Two types
a) Isolated dextrocardia
b) Dextrocardia with situs inversus

Course of the Circulation

No alteration in hemodynamics unless an associated malformation is present

Clinical Features

Asymptomatic
Apex impulse visible on the right
Heart tones heard best over right anterior chest
Liver edge may be palpable in left upper quadrant

Electrocardiogram

Lead I mirror image of normal
Lead II resembles normal lead III
Lead III resembles normal lead II

Radiological Studies

Mirror image of normal
Fluoroscopy of value in establishing position of liver and stomach

Prognosis

a) Isolated dextrocardia—guarded
b) Dextrocardia with situs inversus—good

Treatment

No treatment required unless an associated malformation is present

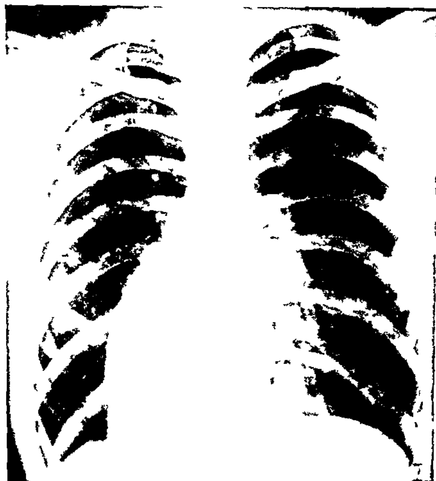


Fig 1

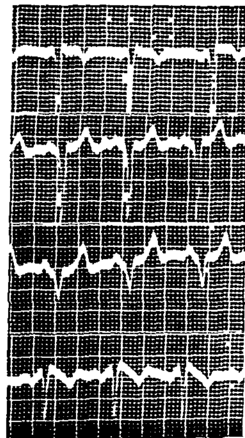


Fig 2

Isolated Dextrocardia

(Adult)

Fig 1 A P roentgenogram of the chest showing typical dextrocardia

Fig 2 E K G Notice that lead I is a mirror image of the normal tracing The Q R S complexe P waves and T waves are inverted in lead I The P waves and T waves are upright in lead III



Fig 3   Fig 4

Tetralogy of Fallot and Situs Inversus

(Age 5 years)

Fig 3 A P roentgenogram of the chest showing dextrocardia associated with tetralogy of Fallot

Fig 4 Heart and lungs Notice the aortic arch curving to the right the left-sided position of the anatomical right ventricle and the large ventricular septal defect

Fig 5 Heart and lungs from the same case Notice the small pulmonary artery to the right of the aorta

Fig 6 E K G Notice that the changes represent a composite of those produced by dextrocardia and the ones due to tetralogy of Fallot The Q R S complexes are upright in lead I and inverted in lead III The P waves and T waves are inverted in lead I and upright in leads II and III The P waves are peaked in lead II



Fig 5

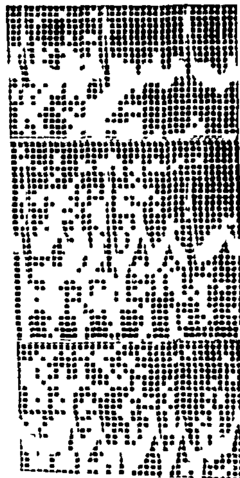


Fig 6

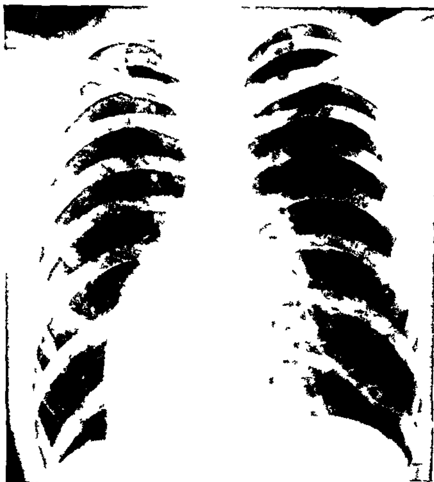


Fig 1

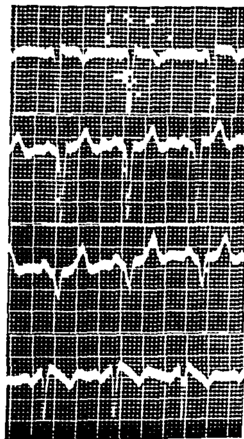


Fig 2

Isolated Dextrocardia

(Adult)

Fig 1 A P roentgenogram of the chest showing typical dextrocardia

Fig 2 E K G Notice that lead I is a mirror image of the normal tracing The Q R S complexes P waves and T waves are inverted in lead I The P waves and T waves are upright in lead III

2 Defects of the Cardiac Septa

Pathology

Defects of the auricular septum include
- a) Persistent foramen ovale
- b) Defects at the site of the fossa ovalis
- c) Defects in the region of the ostium primum
- d) Complete absence of the auricular septum

Defects of the ventricular septum
- a) Defects in the upper membranous septum
- b) Defects in the muscular septum
- c) Complete absence of the ventricular septum

Lutembacher's Syndrome consists of an auricular septal defect associated with a mitral valve stenosis (Plate 1.)

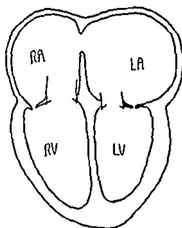


PLATE 1
Lutembacher's syndrome (auricular septal defect associated with mitral valve stenosis)

Course of the Circulation

- a) Patent foramen ovale—right to left shunt only
- b) Other auricular defects—left to right shunt—reversing at times of stress
- c) Ventricular septal defects—left to right shunt—reversing if pulmonary hypertension develops
- d) Lutembacher's Syndrome—usually a large left to right shunt resulting in right sided cardiac enlargement and enlargement of pulmonary artery

Defects of the Auricular Septum

Clinical Features

Poor physical growth and development
Transitory cyanosis in infancy
Repeated respiratory infections
Cardiac enlargement
Narrow pulse pressure
Systolic murmur

2 Defects of the Cardiac Septa

Pathology

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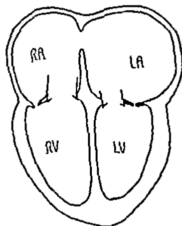


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Defects of the Auricular Septum

Clinical Features

Poor physical growth and development
Transitory cyanosis in infancy
Repeated respiratory infections
Cardiac enlargement
Narrow pulse pressure
Systolic murmur

Electrocardiogram

Right axis deviation
Right ventricular preponderance
Peaked P waves
Prolonged P R interval

Radiological Studies

Right auricular and right ventricular enlargement
Prominent pulmonary artery segment
Increased vascular lung markings
Hilar pulsations noted on fluoroscopy

Laboratory

Microcytic anemia
Increased right auricular pressure
Increased O saturation in right auricle

Prognosis

Poor, may reach adult life but physical activity is usually restricted

Treatment

Prevention of respiratory infection
Treatment of cardiac failure
Surgical closure of defects possible but not completely evaluated



Fig 7

Auricular Septal Defect

Fig 7 Large auricular septal defect in an adult female

Fig 8 A P roentgenogram of the chest in a two-and a half year old child with a large auricular septal defect There is cardiac enlargement and heavy vascular markings in both lung fields



Fig 8

Lutembacher's Syndrome

Except for the huge size of the pulmonary artery on the roentgenograms the clinical features of a Lutembachers' Syndrome are quite similar to those of an auricular septal defect



Fig 9

Lutembacher's Syndrome

(*Age 12 years*)

Fig 9 Heart from a case of Lutembacher's syndrome showing a large auricular septal defect (ostium primum) and a thickened and deformed mitral valve

Fig 10 A P roentgenogram of the chest The picture reveals a large pulmonary conus and heavy vascular lung markings

Fig 11 E K G showing right axis deviation and delayed A V conduction The Q R S complexes are inverted in lead I and upright in lead II The P R interval is 0 20 sec



Fig 10

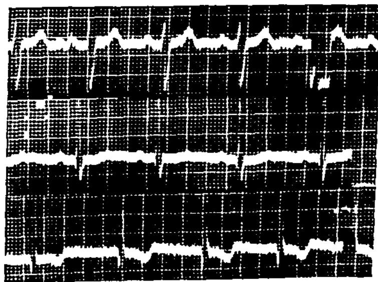


Fig 11



Fig 12

Lutembacher's Syndrome

(Age 7 years)

Fig 12 A P roentgenogram of the chest from another case of Lutembacher's syndrome The picture is similar to Fig 10 but shows a larger right atrium

Fig 13 L A O roentgenogram of the chest from the same case Notice the very prominent right ventricle

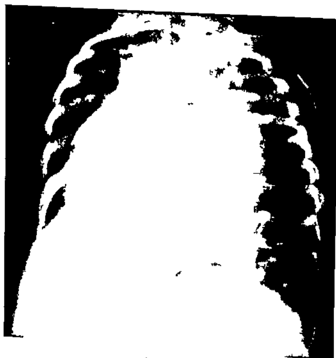


Fig 13

Defects of the Ventricular Septum

Clinical Features

Usually asymptomatic
Growth and development normal
Heart normal size
Systolic murmur—3rd and 4th left I C S close to sternum
Systolic thrill—3rd and 4th left I C S close to sternum

Electrocardiogram

Usually normal

Radiological Studies

Heart normal in size and configuration

Laboratory

O saturation of blood in the right ventricular outflow tract exceeds that of the right ventricle or right auricle

Prognosis

Excellent, unless pulmonary hypertension develops

Treatment

Surgical closure possible but not completely evaluated

Ventricular Septal Defect

Fig 14 Defect in the membranous portion of the ventricular septum The anomaly was found incidentally at autopsy in an adult male

Fig 15 Defect in the muscular portion of the ventricular septum in an infant

Ventricular Septal Defect with Deformity of Aortic Valve

(8 year old male)

Fig 16 Opened left ventricle showing the ventricular septal defect, the deformity of the right aortic leaflet which overlies the septal defect and bacterial endocarditis involving the upper septum and the aortic valve

Fig 17 Close up of the aortic valve and the ventricular septal defect Patient had a continuous murmur at the base of the heart and high pulse pressure

Fig 18 Opened right ventricle showing hypertrophy and ventricular septal defect

Fig 19 A P radiograph of the chest showing cardiac hypertrophy prominence of the pulmonary artery and increased pulmonary vascular markings



Fig 14



Fig 15



Fig 16



Fig 17

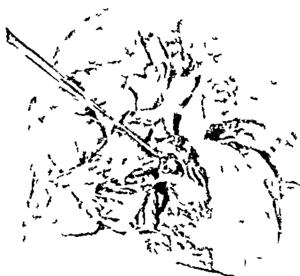


Fig 18



Fig 19

Common Atrioventricular Ostium

Pathology

Incomplete septation of the atrioventricular canal resulting in

a) Defect in the lower portion of the auricular septum
b) Defect in the upper portion of the ventricular septum
c) Common atrioventricular valve

Course of the Circulation

Complete pooling of blood

Clinical Features

Frequently associated with mongolism
Cyanosis slight
Cardiac enlargement
Systolic murmur
Systolic thrill

Electrocardiography

No diagnostic pattern
Usually right axis deviation

Radiological Studies

Cardiac enlargement
Increased vascular lung markings

Laboratory

Decreased arterial O saturation

Prognosis

Average length of life ten to twelve years

Treatment

Prevention of respiratory infections
Management of cardiac failure



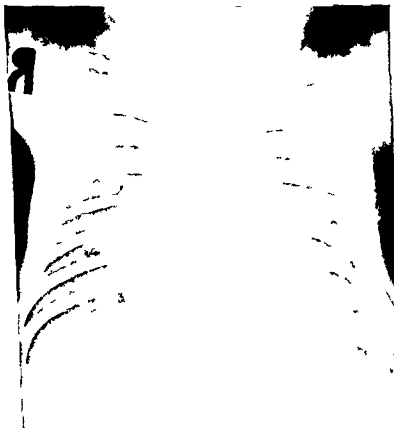
Fig 20

Common Atrioventricular Ostium

(6 month old male)

Fig 20 Common atrioventricular ostium Notice the opened right ventricle the large defect in the lower part of the auricular septum and the common valve with leaflets crossing the midline through the septal defect

Fig 21 A P roentgenogram of the chest from the same case as the specimen in Fig 20 The heart is enlarged to right and left and the pulmonary vascular markings are heavy



Common Atrioventricular Ostium

Pathology

Incomplete septation of the atrioventricular canal resulting in

a) Defect in the lower portion of the auricular septum
b) Defect in the upper portion of the ventricular septum
c) Common atrioventricular valve

Course of the Circulation

Complete pooling of blood

Clinical Features

Frequently associated with mongolism
Cyanosis slight
Cardiac enlargement
Systolic murmur
Systolic thrill

Electrocardiography

No diagnostic pattern
Usually right axis deviation

Radiological Studies

Cardiac enlargement
Increased vascular lung markings

Laboratory

Decreased arterial O saturation

Prognosis

Average length of life ten to twelve years

Treatment

Prevention of respiratory infections
Management of cardiac failure



Fig 22



Fig 23



Fig 24

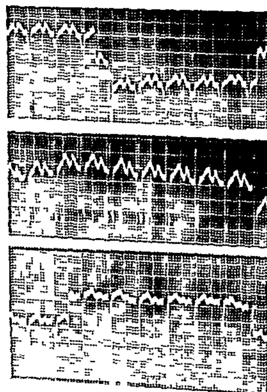


Fig 25

Common Atrioventricular Ostium

(16 month old male)

Fig 22 Heart from a boy with common atrioventricular ostium Picture shows the left atrium and left ventricle with a large septal defect Notice the common A V valve with some leaflets running through the septal defect

Fig 23 Common atrioventricular orifice with six valvular leaflets two common leaflets (across midline) one leaflet in the left ventricle, and three in the right ventricle (two of these are rudimentary)

Fig 24 A P roentgenogram of chest from the same case shows marked cardiac enlargement and congestion in the lung fields

Fig 25 E K G showing tachycardia prominence of P_1 and P and a diphasic P_3 also right axis deviation

Cor Triloculare

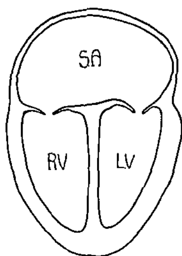


PLATE 3
Cor triloculare biventriculare (a three chamber heart a single auricle and two ventricles)

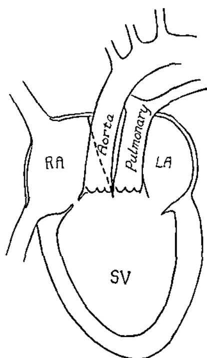


PLATE 4
Cor triloculare biatriatum (a three chamber heart with two auricles and a single ventricle

Pathology (Plates 3 4)

Complete absence of auricular septum—*cor triloculare biventriculare*
Complete absence of ventricular septum—*cor triloculare biatriatum*
There are usually other associated cardiovascular anomalies

Course of the Circulation

Complete pooling within the common chamber

Clinical Features

Cyanosis from early infancy
Cardiac enlargement
Poor growth and development
Systolic murmur

Electrocardiogram

Right axis deviation
Abnormal P waves
Delayed A V conduction

Radiological Studies

Cardiac enlargement
Lung fields appear congested

Laboratory

Arterial O_2 saturation decreased

Prognosis

Poor usually expire in early infancy

Treatment

None

Cor Biloculare

Pathology (Plate 2)

Complete absence of auricular and ventricular septa
Common atrioventricular ostium
Occasionally associated lesions are transposition of the great vessels and truncus arteriosus

Course of the Circulation

Complete pooling within the common auricle and ventricle

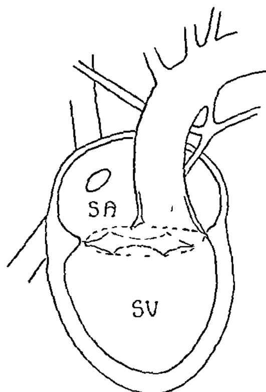


PLATE 2
Cor biloculare (a two chamber heart, a single auricle and single ventricle)



Fig 26

Cor Biloculare

Fig 26 Heart specimen from an infant with cor biloculare Notice the complete absence of auricular and ventricular septa and the common atrioventricular valve

Cor Triloculare

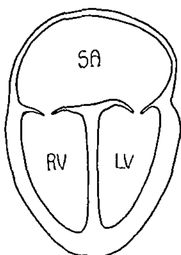


PLATE 3
Cor triloculare biventriculare (a three chamber heart a single auricle and two ventricles)

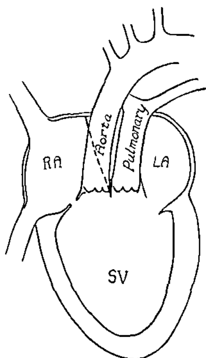


PLATE 4
Cor triloculare biatriatum (a three chamber heart with two auricles and a single ventricle

Pathology (Plates 3 4)

Complete absence of auricular septum—*cor triloculare biventriculare*
Complete absence of ventricular septum—*cor triloculare biatriatum*
There are usually other associated cardiovascular anomalies

Course of the Circulation

Complete pooling within the common chamber

Clinical Features

Cyanosis from early infancy
Cardiac enlargement
Poor growth and development
Systolic murmur

Electrocardiogram

Right axis deviation
Abnormal P waves
Delayed A V conduction

Radiological Studies

Cardiac enlargement
Lung fields appear congested

Laboratory

Arterial O_2 saturation decreased

Prognosis

Poor usually expire in early infancy

Treatment

None



Fig. 27



Fig. 28

Cor Triloculare Biventriculare

(Age 4 months)

Fig. 27. Heart from a case of cor triloculare biventriculare. Notice the complete absence of auricular septum, and the small ventricular septal defect. Notice also the common atrioventricular valve.

Fig. 28. A.P. roentgenogram from same case of cor *triloculare biventriculare as shown in Fig. 27.* There is a large globular heart and very vascular lung fields.



Fig 29

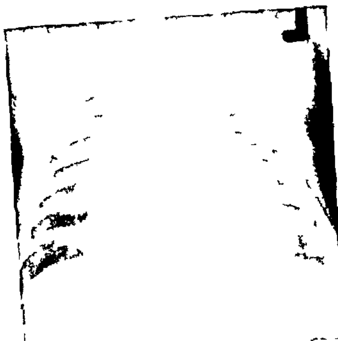


Fig 30

Cor Triloculare Biatriatum

(*Age 8 days*)

Fig 29 Heart from a case of cor triloculare biatriatum Notice the single ventricle with the four cardiac valves opening into it Fig 30 A-P roentgenogram from same case of cor triloculare biatriatum as shown in Fig 29 It shows cardiac enlargement and increased pulmonary vascular markings Fig 31 Section of lung from same case of cor triloculare biatriatum Notice the thick walled blood vessels especially in the subpleural region



Fig 31



Fig 32



Fig 33

Cor Triatriatum

(6 months old female)

Fig 32 Superior view of the heart showing the accessory left atrium The probes mark the points of entry of all four pulmonary veins A short probe is directed down toward the main left atrium

Fig 33 Left ventricle mitral valve and left atrium The only communication between the left atrium and the accessory atrium is through a narrow opening as shown by probe

3 Valvular Anomalies

Tricuspid Valve Atresia

Pathology (Plate 5)

Complete obliteration of tricuspid orifice
Hypoplasia of right ventricle
Hypoplasia of pulmonary valve and artery
Associated defects of the auricular and ventricular septa
Patent ductus arteriosus

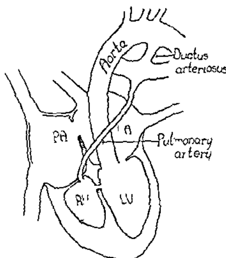


PLATE 5
Tricuspid atresia with under developed right ventricle

Course of the Circulation

Right auricle → left auricle → left ventricle → aorta
→ systemic circulation
↘ patent ductus or bronchial arteries to the lung

Clinical Features

Cyanosis from birth a common finding
Heart normal size
Systolic murmur
Enlarged liver
Pulsations at liver margin present on occasion

Electrocardiogram

Usually left axis deviation
Left ventricular preponderance
Peaked P waves
(Fig 35 an exception)

Radiological Findings

Decreased bronchovascular markings
Heart size normal
Concavity in the region of the pulmonary conus P A view
Right cardiac border has double convexity P A view
Shadow cast by right ventricle absent in L A O view

Laboratory

Arterial O_2 unsaturation
Hemoconcentration

Prognosis

Poor
Average length of life 4–6 months

Treatment

Surgical creation of an auricular defect or the creation of an extracardiac shunt is of benefit

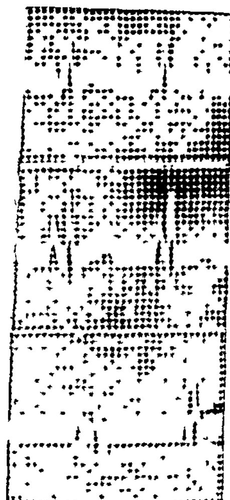


Fig 35



Fig 34

Tricuspid Atresia and Pulmonary Valve Atresia

(Age 4 months)

Fig 34 Heart in a case of tricuspid atresia Notice the dilated right atrium and the atretic tricuspid orifice

Fig 35 E K G showing peaked P wave in leads II and III but no evidence of left axis deviation

Fig 36 Pulmonary artery and valve in the same case as Fig 34 Notice the atretic pulmonary valve orifice

Fig 37 A P roentgenogram of chest in the same case The heart is of normal size and shows slight concavity in the region of the pulmonary conus and a double convexity of the right cardiac border The lung fields are avascular

Fig 38 L A O roentgenogram of chest Notice the normal size of the left *(posterior)* ventricle and the small size of the right *(anterior)* ventricle



Fig 36



Fig 37



Fig 38



Fig 39



Fig 40

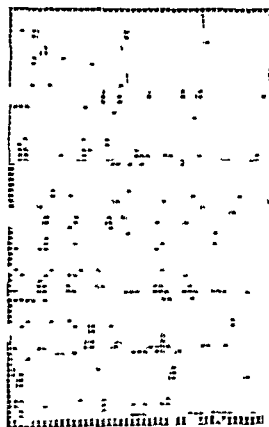


Fig 41

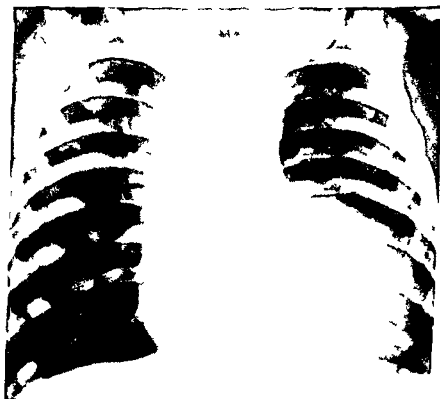


Fig 42

Tricuspid Atresia
(with absence of auricular septum)

(Age 2½ years)

Fig 39 Heart from male with tricuspid atresia Notice the absence of auricular septum and the atretic tricuspid orifice

Fig 40 Anterior view of the same heart as in Fig 39 Notice the small pulmonary artery and the calcified nodular thrombus in the infundibulum

Fig 41 E K G Notice the peaked P waves in leads I and II with evidence of left axis deviation The Q R S complexes are upright in lead I and inverted in lead III

Fig 42 A P roentgenogram of the chest from a 2½ year old male with tricuspid atresia and complete absence of auricular septum Notice the left sided enlargement of the heart the concavity in the region of the pulmonary conus and the double convexity in the right border The lung fields are avascular

Fig 43 Sections of aorta and pulmonary artery The longer section is from the pulmonary artery At 2½ years the two sections should be of about equal thickness Elastic tissue stain (× 15)

Fig 44 Section of lung Notice the thin walled pulmonary arteries and their relatively small size Elastic tissue stain (× 15)

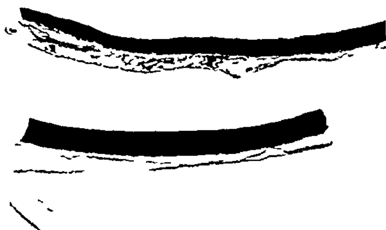


Fig 43



Fig 44

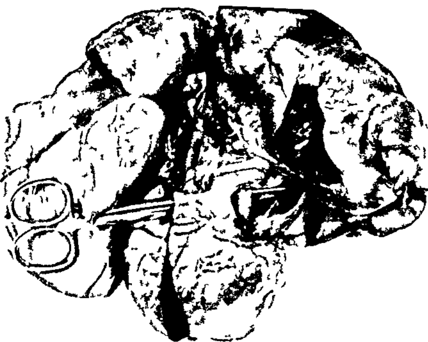


Fig 45



Fig 46



Fig 47



Fig 48



Fig 49

Tricuspid Atresia
(with auricular septal defect)

(Age 10 years)

Fig 45 Heart and lungs from a 10 year old colored male with tricuspid atresia The scissors pass through a large defect in the auricular septum Notice the atresia of the tricuspid orifice

Fig 46 Anterior view of the same heart Notice the prominent left ventricle the small pulmonary artery and the undeveloped right ventricle which consists only of a small outflow (infundibular) portion The probe passes through a small ventricular septal defect

Fig 47 View of the heart showing small hypoplastic pulmonary artery

Fig 48 Descending thoracic aorta *showing the intercostal arteries and* some large bronchial arteries

Fig 49 Section of the aorta and pulmonary artery Notice the thin walled pulmonary artery which is the longer of the two specimens shown

Fig 50 Section of the lung showing small and thin walled pulmonary arteries Elastic tissue stain (× 15)

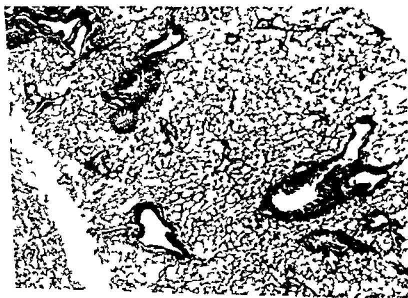


Fig 50

Pulmonary Valve Atresia

Pathology (Plate 6)

Pulmonary valve obliterated
Pulmonary artery reduced in size
A *patent ductus arteriosus frequently* present
Auricular septal defect
Large dilated right auricle
Small and thick walled right ventricle

Course of the Circulation

Right auricle → left auricle → left ventricle
aorta ——→ systemic circulation
↘ patent ductus or enlarged bronchial arteries to the lungs

Clinical Features

Cyanosis and dyspnea
Progressive cardiac enlargement
Liver enlarged

Electrocardiography

Right axis deviation
Peaked P waves are common

Radiological Studies

Marked cardiac enlargement (right auricle)
Avascularity of lung fields

Laboratory

Arterial O unsaturation
Hemoconcentration
Increased right ventricular and right auricular pressure revealed by catheterization

Prognosis

Poor
Death at 4–6 months

Treatment

Pulmonary valvulotomy of value if the pulmonary artery is not markedly hypoplastic

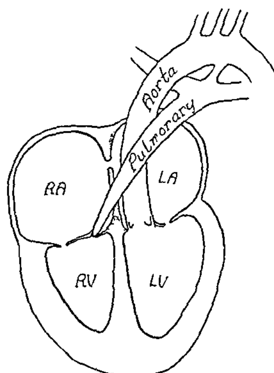


PLATE 6
Pulmonary valve atresia

Pulmonary Valve Stenosis

Pathology

Fusion of valvular cusps into a small "volcano-like" structure with the narrow valvular orifice as the crater
Post-stenotic dilation of the pulmonary artery
Right ventricular hypertrophy
Often an associated auricular septal defect or patent foramen ovale
Intact ventricular septum

Course of the Circulation

In most instances the hypertrophic right ventricle forces blood through the stenotic orifice
If an auricular septal defect is present the flow is from right to left

Clinical Features

Variable depending upon absence or presence of auricular defect
Dyspnea an outstanding symptom
Cyanosis if auricular defect is present
Heart normal in size
Systolic murmur
Systolic thrill
Decreased or absent P

Electrocardiogram

Right axis deviation
Right ventricular preponderance
Huge peaked P waves

Radiological Studies

Heart normal size or slightly enlarged
Vascular lung markings normal or decreased
Prominent pulmonary conus (post-stenotic dilation)
Angiocardiography of value in demonstrating presence of an auricular septal defect

Laboratory

There is usually arterial O_2 unsaturation
Catheterization reveals an increased right ventricular pressure and a normal or low pulmonary artery pressure

Prognosis

In general good

Treatment

Pulmonary valvulotomy

Pulmonary Valve Atresia

Pathology (Plate 6)

Pulmonary valve obliterated
Pulmonary artery reduced in size
A patent ductus arteriosus frequently present
Auricular septal defect
Large dilated right auricle
Small and thick walled right ventricle

Course of the Circulation

Right auricle → left auricle → left ventricle
aorta ⟶ systemic circulation
↘ patent ductus or enlarged bronchial arteries to the lungs

Clinical Features

Cyanosis and dyspnea
Progressive cardiac enlargement
Liver enlarged

Electrocardiography

Right axis deviation
Peaked P waves are common

Radiological Studies

Marked cardiac enlargement (right auricle)
Avascularity of lung fields

Laboratory

Arterial O unsaturation
Hemoconcentration
Increased right ventricular and right auricular pressure revealed by catheterization

Prognosis

Poor
Death at 4–6 months

Treatment

Pulmonary valvulotomy of value if the pulmonary artery is not markedly hypoplastic

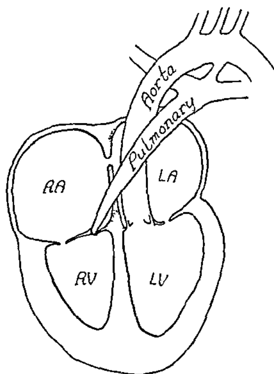


Plate 6
Pulmonary valve atresia



Fig 53

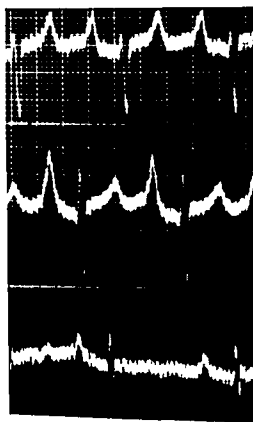


Fig 54



Fig 51



Fig 52

Pulmonary Valve Atresia

(*Age 8 weeks*)

Fig 51 Pulmonary valve atresia Notice the membranous formation which obliterates the pulmonary valvular orifice The right ventricle is small but very thick walled The right atrium is enormously dilated The ductus arteriosus is patent

Fig 52 Posterior view of the heart Notice the dilated right atrium the small left atrium, and the auricular septal defect

Fig 53 A P roentgenogram of the chest Notice the marked cardiac enlargement to the right and left The large right atrium can be seen

Fig 54 E K G showing peaked P waves in leads I and II There is right axis deviation The Q R S complexes are inverted in lead I and upright in lead III

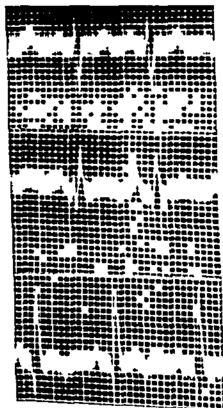


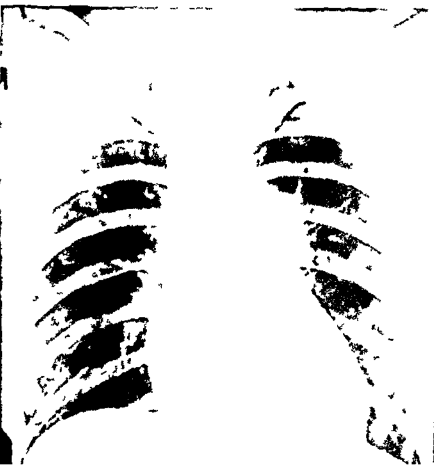
Fig 57



Fig 58



Fig 55



Pulmonary Valve Stenosis
(with auricular septal defect)

(25 year old female)

Fig 55 Pulmonary valve stenosis Pulmonary valve seen from above Notice the small circular opening and the position of the commissures Notice also the marked dilatation of the pulmonary artery

Fig 56 A P roentgenogram of the chest Notice the large pulmonary artery and the avascular lung fields

Fig 57 E K G showing a marked right axis deviation and peaked P waves in lead II The Q R S complexes are inverted in lead I and upright in lead III

Fig 58 Sections of aorta and pulmonary artery Notice the extremely thin wall of the pulmonary artery Elastic tissue stain (X 15)



Fig 62

Pulmonary Infundibular Stenosis

(69 year old female)

Fig 62 Notice the narrow opening in the outflow tract of the right ventricle The ventricular septum is intact The pulmonary valve is unchanged



Fig 59

Pulmonary Valve Stenosis
(with an intact auricular septum)

(*Age 8 months*)

Fig 59 Heart and lungs from an 8 month old child with pulmonary valve stenosis Notice the typical deformity of the pulmonary valve with a round crater in the center There is moderate pulmonary dilatation, and marked hypertrophy of the right ventricle and right atrium

Fig 60 Section of aorta and pulmonary artery Notice the marked thinness of the pulmonary artery wall Elastic tissue stain (× 15)

Fig 61 Section of lung revealing small and thin walled pulmonary arteries Elastic tissue stain (× 15)

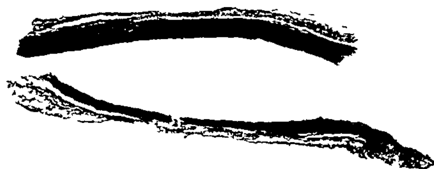


Fig 60



Aortic Valve Atresia

Pathology (Plate 8)

Aortic valve atresia
Hypoplasia of left ventricle
Hypoplastic aorta
Right auricular right ventricular enlargement
Pulmonary artery enlarged
Patent ductus arteriosus

Course of the Circulation

Left ventricular and left auricular blood → right auricle → right ventricle → pulmonary artery → patent ductus arteriosus → aorta

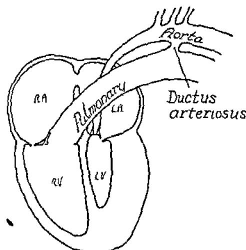


PLATE 8
Aortic valve atresia

Clinical Features

Progressive cyanosis and cardiac enlargement
Systolic murmur

Electrocardiogram

Right axis deviation
Peaked P waves

Radiological Studies

Cardiac enlargement
Normal or increased vascular lung markings

Laboratory

Arterial O_2 saturation is decreased
Hemoconcentration
O_2 content in right auricular blood sample increased

Prognosis

Poor
Most infants with this malformation die within the first few weeks of life

Treatment

None

Mitral Valve Atresia

Pathology (Plate 7)

Atresia of the mitral valve
No communication between left auricle and ventricle
Patent foramen ovale or auricular septal defect
Hypoplastic left ventricle
Ventricular septal defect
Hypoplastic aorta and aortic valve

Course of the Circulation

Left auricle → right auricle → right ventricle
right ventricle → pulmonary artery
↘ ventricular septal defect → aorta

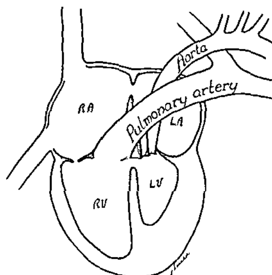


Plate 7
Mitral valve atresia

Clinical Features

Progressive cyanosis from early infancy
Early cardiac enlargement
Systolic murmur

Electrocardiogram

Right axis deviation is the usual finding

Radiological Studies

Marked cardiac enlargement
Prominent pulmonary artery
Angiocardiograms may reveal simultaneous filling of the aorta and pulmonary artery

Laboratory

Arterial O_2 unsaturation
O_2 content of right auricular blood increased

Prognosis

Poor
Death in early infancy

Treatment

None

4 Anomalies of Septation of Truncus Arteriosus

Persistent Truncus Arteriosus

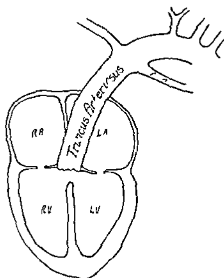


PLATE 9
Complete truncus arteriosus

Pathology

There is only one vessel arising from the base of the heart. This single vessel is the origin of the coronary, pulmonary, and systemic vessels.

A defect in the membranous portion of the ventricular septum.

There are frequently four cusps in the valve of the vessel arising from the base of the heart.

Plates 9 and 10 illustrate complete and partial truncus arteriosus.

Course of the Circulation

The ventricular septal defect and the common trunk for the pulmonary and systemic circulations result in marked pooling of arterial and venous blood.

Circulation to the lung is by way of pulmonary vessels arising from the truncus or by the way of bronchial vessels.

Clinical Features

Cyanosis moderate
Heart normal size or slightly enlarged
Systolic murmur
Clear cut second sound

Electrocardiogram

Right axis deviation



Fig 63



Fig 64

Aortic Valve Atresia

(6 months old male)

Fig 63 Large and thick walled pulmonary artery hypoplastic aorta and patent ductus arteriosus Notice the large right atrium and right ventricle

Fig 64 Posterior view of the heart showing marked hypoplasia of the left atrium and left ventricle, a patent foramen ovale and *hypertrophy and dilatation of both right sided chambers*

Persistent Truncus Arteriosus Partial Type

(Age 8 days)

Fig 65 Partial truncus arteriosus communis persistens showing incomplete aortic pulmonary septum left coronary ostium and short pulmonary trunk with its two branches

Fig 66 A P roentgenogram of the chest Notice marked enlargement of cardiac outline to left absence of pulmonary conus and small size of aortic knob

Fig 67 L A O roentgenogram of the chest Notice the large left ventricle The anterior border of the right ventricle is only slightly more prominent than the border of the common trunk



Fig 65

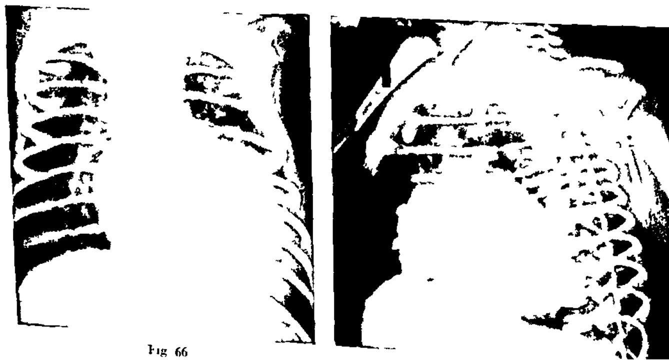


Fig 66

Radiological Studies

Right and left ventricular enlargement
When cyanosis is minimal, marked hilar pulsations are usually present
When cyanosis is marked the lung fields are avascular

Laboratory

O saturation of the arterial blood decreased
Hemoconcentration may be present

Prognosis

Variable—length of life depends on blood supply to the lungs

Treatment

None

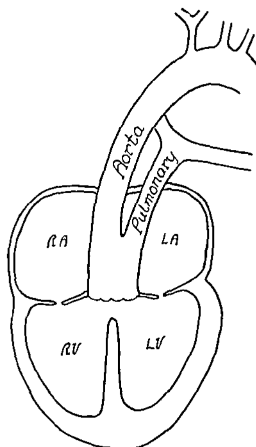
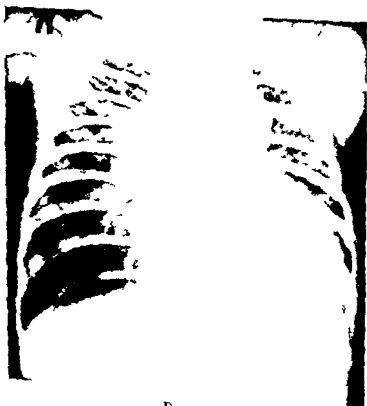


PLATE 10
Partial truncus arteriosus



Fig 71



F

**Persistent Truncus Arteriosus
Complete Type**

(*Age 10 months*)

Fig 71 Complete truncus *arteriosus communis* Notice displacement of arterial trunk to the right four fleshy *cusps* single large coronary ostium and large pulmonary arteries coming directly off the truncus

Fig 72 A P roentgenogram of the *chest* Notice marked enlargement of *cardiac* shadow to left absence of pulmonary conus and enormous aortic knob



Fig 68

Persistent Truncus Arteriosus, Partial Type

(Age 6 weeks)

Fig 68 Partial truncus arteriosus communis persistens Notice incomplete aortic pulmonary septum between pulmonary trunk and ascending aorta The ventricular septal defect is plainly seen

Fig *69* A P roentgenogram of the chest Notice the marked enlargement of the cardiac outline to the left and the absence of the pulmonary conus, also the small size of the aortic knob

Fig 70 L A O roentgenogram of the chest Notice the large left ventricle The anterior border of the right ventricle is only slightly more prominent than the border of the common trunk



Fig 69



Fig 70

Solitary Aortic and Pulmonic Trunk

Pathology (Plates 11, 12)

Eccentric septation of the truncus arteriosus
Solitary aortic trunk with pulmonary atresia—or
Solitary pulmonic trunk with aortic atresia
Ventricular septal defect

Course of the Circulation

Solitary aortic trunk—blood from both ventricles enters the solitary arterial trunk and the blood reaches the lungs through a patent ductus

Solitary pulmonic trunk—blood from both ventricles enters the pulmonic trunk, pulmonary vessels arise from the pulmonic trunk and the ductus arteriosus continues as the descending aorta

Clinical Features

Similar to that of a truncus arteriosus or partial truncus arteriosus

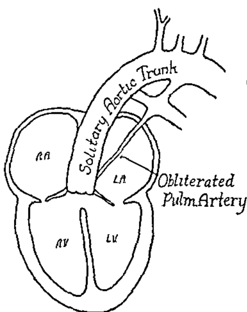


Plate 11
Solitary aortic trunk

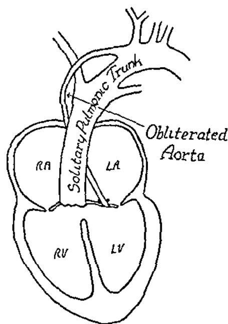


Plate 12
Solitary pulmonic trunk



Fig 73

Persistent Truncus Arteriosus, Partial Type

(*Age 7 days*)

Fig 73 "Partial" truncus arteriosus communis Notice the four "fleshy" cusps in the valve, the incomplete aortic pulmonary septum, and the ventricular septal defect



Persistent Truncus Arteriosus with Right Aortic Arch and Right Descending Aorta

(*Age 3 weeks*)

Fig 74 "Partial" truncus arteriosus communis Notice the incomplete aortic pulmonary septum and the right sided aortic arch

Aortic Septal Defect

Pathology (Plate 13)

Incomplete septation of the truncus arteriosus
Both great vessels arise from the heart in a normal fashion
Communication between pulmonary artery and aorta close to the base of the heart

Course of the Circulation

Flow through aortic septal defect is left to right, that is from aorta to pulmonary artery

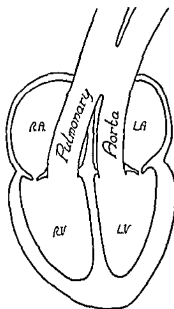


PLATE 13
Aortic septal defect

Clinical Features

Growth and development may be retarded or normal
Peripheral signs of aortic insufficiency
Continuous roaring murmur over the base of the heart
Thrill over base of heart

Electrocardiogram

Usually normal
On occasion left ventricular preponderance

Radiological Studies

Vascular lung markings increased
Hilar pulsations observed on fluoroscopy
Evidence of left ventricular enlargement

Laboratory

Arterial O_2 saturation normal
Catheterization reveals O_2 content of blood from pulmonary artery exceeds that of the right ventricle
Pulmonary pressure may be increased

Prognosis

Good

Treatment

Surgical closure of the defect



Fig 75

Solitary Aortic Trunk with Pulmonic Atresia

(Age 8 days)

Fig 75 Heart from a case of solitary aortic trunk (pseudotruncus?) Notice the large aorta and the atretic pulmonary artery Notice also the patent ductus arteriosus which communicates with the arterial branches to both lungs

5 Transposition Complexes

Eisenmenger's Complex

Pathology (Plate 14)

Right ventricular hypertrophy
Ventricular septal defect
Dextroposition of the aorta
Pulmonary artery large and dilated

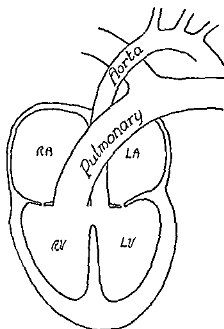


PLATE 14
Eisenmenger's complex

Course of the Circulation

The dextroposed aorta receives blood from both ventricles
The enlarged pulmonary artery results in an increased flow and increased pressure in the pulmonary circulation

Clinical Features

Onset of cyanosis in late childhood
Slight cardiac enlargement
Basal systolic murmur and thrill
Hemoptysis is frequent

Electrocardiogram

Right axis deviation
Right ventricular preponderance
Peaked P waves

Radiological Studies

Contour of the heart varies with the size of the pulmonary artery
If pulmonary artery is enlarged contour resembles that seen in cases of Lutembacher's syndrome
If pulmonary artery is not enlarged, contour resembles that of a Tetralogy of Fallot

Radiological Studies

Heart normal size
Concave pulmonary conus
Upturned apex
25% right aortic arch
Avascular lung fields
Angiocardiography of value in determining position of aorta

Laboratory

Arterial O saturation markedly reduced
Hemoconcentration
Cardiac catheterization reveals increased right ventricular pressure catheter may pass through ventricular defect into aorta

Prognosis

Compatible with long life however physical activity may be moderately or markedly restricted

Treatment

Surgical designed to increase blood flow to lungs
Blalock Taussig subclavian artery pulmonary artery anastomosis
Potts Smith direct aorta pulmonary artery anastomosis
Brock Procedure direct attack on the pulmonary stenosis

The lung fields are quite vascular, hilar pulsations conspicuous
Angiocardiography results in simultaneous filling of the right ventricle, enlarged pulmonary artery and aorta

Laboratory

Arterial O_2 unsaturation
Catheterization reveals an increased right ventricular and pulmonary artery pressure

Prognosis

Once cyanosis develops the prognosis is guarded

Treatment

No surgical treatment

Tetralogy of Fallot

(Illustrations begin page 56)

Pathology (Plate 15)

Pulmonary stenosis valvular or infundibular
Dextroposition of the aorta
Right ventricular hypertrophy
Ventricular septal defect

Course of the Circulation

The dextroposed aorta receives blood from both ventricles
Decreased pulmonary flow as a result of the pulmonary stenosis
Bronchial arteries become important in blood supply to lungs

Clinical Features

Cyanosis and clubbing
Heart normal size
Left anterior chest deformity
Basal systolic murmur and thrill

Electrocardiogram

Right axis deviation
Right ventricular preponderance
Peaked P waves

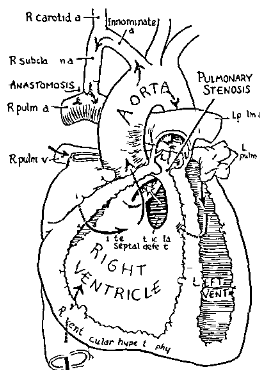


Plate 15
Tetralogy of Fallot with a Blalock Taussig shunt

Eisenmenger Complex
(with right sided aortic arch and descending aorta)

(Age 8 months)

Fig 76 Eisenmenger complex Notice the large size of the pulmonary artery The arch of the aorta and the descending portion are to the right of the trachea

Fig 77 Eisenmenger complex Same case showing the opened left ventricle and the high ventricular septal defect

Fig 78 A P roentgenogram of the chest Notice the cardiac enlargement and the heavy vascular markings of the lungs

Fig 79 E K G showing right axis deviation peaked P waves in lead II and diphasic P waves in lead III The Q R S complex is inverted in lead I and upright in lead III

Fig 80 Eisenmenger complex Section of aorta and pulmonary artery (longest section) Notice the hypertrophy of the pulmonary artery which is thicker than the aorta Elastic tissue stain (X 15)

Fig 81 Eisenmenger complex Section of lung showing the large and thick walled pulmonary arteries Elastic tissue stain (X 15)

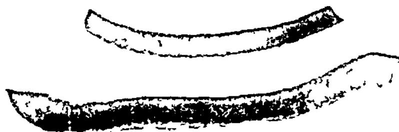


Fig 80

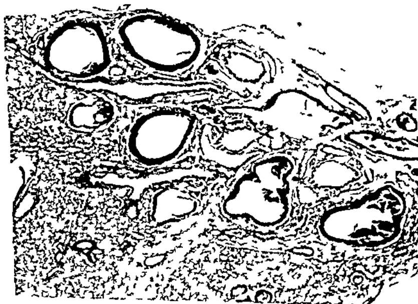


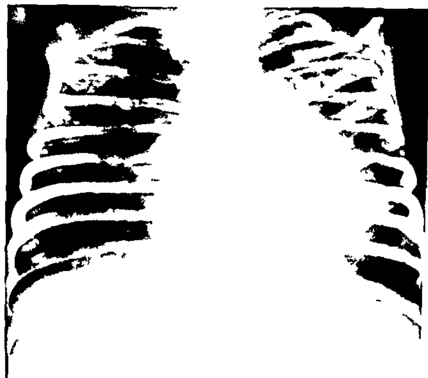
Fig 81

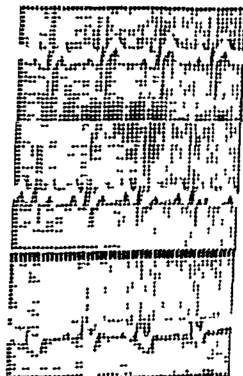


Fig 76



Fig 77





Eisenmenger Complex
(with right sided aortic arch and descending aorta)

(Age 8 months)

Fig 76 Eisenmenger complex Notice the large size of the pulmonary artery The arch of the aorta and the descending portion are to the right of the trachea

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Fig 81 Eisenmenger complex Section of lung showing the large and thick walled pulmonary arteries Elastic tissue stain (X 15)

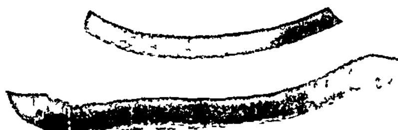


Fig 80

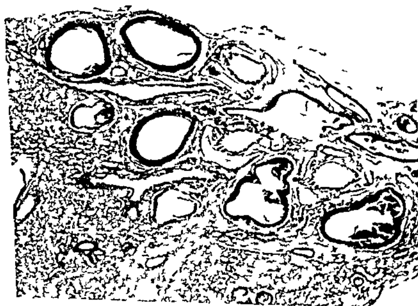


Fig 81

Eisenmenger Complex

(*Age 18 months*)

Fig 82 Eisenmenger complex Notice the large pulmonary artery and the small aorta Notice also the hypertrophy of the right ventricle

Fig 83 Eisenmenger complex The probe passes through the ventricular septal defect

Fig 84 Eisenmenger complex Sections of aorta and pulmonary artery (longest section) Notice the pronounced relative thickness of the pulmonary artery Elastic tissue stain (× 15)

Fig 85 Eisenmenger complex Section of lung showing the large and thick walled pulmonary arteries Elastic tissue stain (× 15)



Fig 82



Fig 83



Fig 84

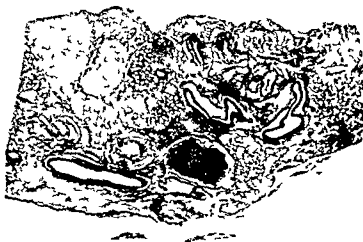


Fig 85



Fig 86

Eisenmenger Complex with Interruption of Aortic Arch

(*Age 6 months*)

Fig 86 Eisenmenger complex with interruption of the aortic arch Notice the hypertrophy of the right ventricle and the pulmonary artery Notice also the interruption of the aortic arch with the pul monary artery running into the descending aorta through a patent ductus arteriosus The right subclavian and right carotid arteries originate proximal to the interruption while the left subclavian and left carotid arteries originate in the region of the patent ductus arteriosus



Fig 87



Fig 88

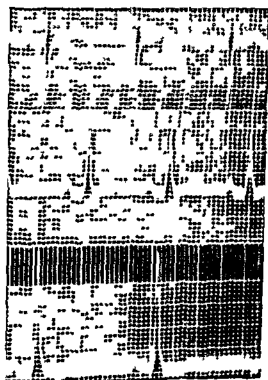


Fig 89



Fig 90



Fig. 91

Tetralogy of Fallot
(after Potts-Smith operation)

(A 17 year old male)

Fig 87 Heart from a seventeen year old male with tetralogy of Fallot Notice the large right ventricle the dextroposed aorta and the large ventricular septal defect Notice also the rudimentary third (infundibular) ventricle from which originates a very small pulmonary artery

Fig 88 A P roentgenogram of the chest The shape of the heart is normal The pulmonary conus is slightly concave and the lung fields are avascular

Fig 89 E K G revealing a very pronounced right axis deviation and peaked P waves in lead II The Q R S complex is inverted in lead I and upright in leads II and III

Fig 90 Descending thoracic aorta showing a probe in the surgically created ductus arteriosus Notice also the large bronchial arteries

Fig 91 Tetralogy of Fallot Sections of aorta and pulmonary artery Notice the very thin pulmonary artery Elastic tissue stain (X 15)

Fig 92 Tetralogy of Fallot Section of lung showing the small size and thin wall of the pulmonary arteries



Fig. 92

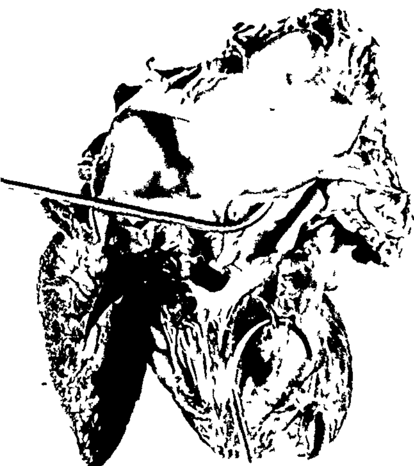


Fig 93

Tetralogy of Fallot

(*Age 6 years*)

Fig 93 Tetralogy of Fallot Notice the large right ventricle the dextroposed aorta the small pulmonary artery, and the ventricular septal defect

Fig 94 A P roentgenogram of the chest Notice the upturned apex and the avascular lung fields

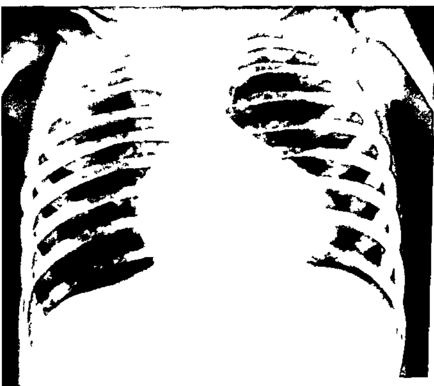


Fig 94



Fig. 95

Tetralogy of Fallot with Bulbar Inversion

(*Age 3½ years female*)

Fig. 95 Tetralogy of Fallot with bulbar inversion Notice the hypertrophy of the right ventricle the dextroposed aorta the large ventricular septal defect and the small pulmonary artery behind the aorta



Fig 96



Fig 97

Tetralogy of Fallot and Dextrocardia

(Age 2 years)

Fig 96 Tetralogy of Fallot and dextrocardia Notice the arch of the aorta to the right of the trachea

Fig 97 Tetralogy of Fallot with dextrocardia Notice the anatomical right atrium with a probe in the superior vena cava, and two other probes in the pulmonary veins of the left lung The other two pulmonary veins drained normally into the anatomical left atrium

Taussig-Bing Heart

Pathology (Plate 16)

Aorta arises from the right ventricle
Pulmonary artery over rides a ventricular septal defect
Right ventricle and pulmonary artery hypertrophic

Course of the Circulation

Pulmonary artery receives blood from both ventricles
Aorta receives blood only from the right ventricle
There is increased flow and pressure in the pulmonary circulation

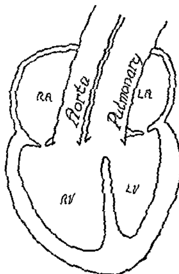


Plate 16
Taussig Bing heart

Clinical Features

Similar to that produced by Eisenmenger's complex, however, in this instance cyanosis is present from birth

Electrocardiogram

Right axis deviation
Right ventricular preponderance

Radiological Studies

Heart moderately enlarged
Pulmonary conus is full
Vascular lung markings increased

Laboratory

Arterial O_2 unsaturation
Hemoconcentration
Catheterization reveals O_2 content of the right ventricular blood sample to be increased. Catheter may enter the aorta directly from the right ventricle

Prognosis

Compatible with life for several years

Treatment

None



Fig 96



Fig 97

Tetralogy of Fallot and Dextrocardia

(Age 2 years)

Fig 96 Tetralogy of Fallot and dextrocardia Notice the arch of the aorta to the right of the trachea

Fig 97 Tetralogy of Fallot with dextrocardia Notice the anatomical right atrium with a probe in the superior vena cava, and two other probes in the pulmonary veins of the left lung The other two pulmonary veins drained normally into the anatomical left atrium



Fig 100

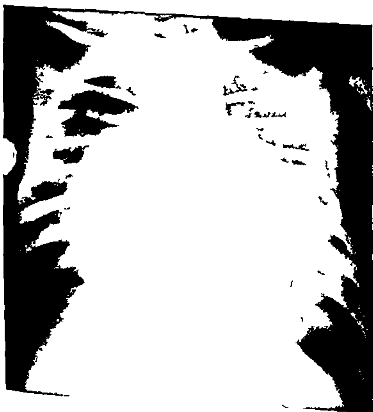


Fig 101

Taussig-Bing Heart

(*Age 7½ months*)

Fig 100 A P roentgenogram of the chest taken shortly after birth Notice the cardiac enlargement especially on the right side

Fig 101 A P roentgenogram of the chest of the same patient at 7½ months of age Notice the enormously dilated globular heart

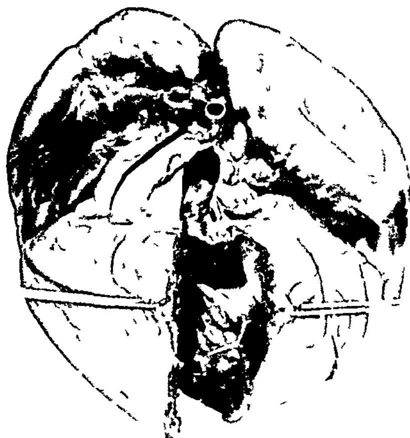


Fig 98

Taussig Bing Heart

(Transposition of the aorta and levoposition of the pulmonary artery)

(*Age 2½ years*)

Fig 98 Heart and lungs from a case of Taussig Bing complex Notice the large defect in the ventricular septum and the pulmonary valve directly above the defect Notice also the completely transposed aorta

Fig 99 A P roentgenogram of the chest Notice the cardiac enlargement to the right and left, and the increased hilar markings



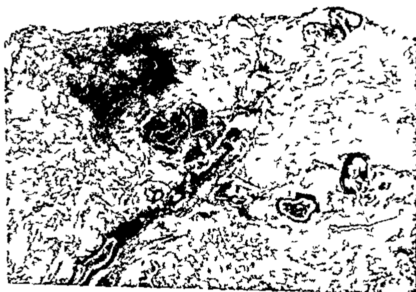


Fig 101



Fig 102



Corrected Transposition of the Aorta and Levoposition of the Pulmonary Artery

(Taussig Bing Heart with Corrected Transposition of the Aorta)

(*Age 1¼ years*)

Fig 102 Heart showing hypertrophy of the right ventricle the anterior position of the aorta, and the large ventricular septal defect below the pulmonary valve

Fig 103 Heart viewed from the left lateral side The transposition is readily shown The ductus arteriosus is patent

Fig 104 Section of lung showing pulmonary arteriosclerosis and necrotizing arteritis The latter process was confined to the pulmonary arteries



Fig. 104

Complete Transposition of the Arterial Trunks

Pathology (Plate 17)

Aorta arises from the right ventricle
Pulmonary artery arises from the left ventricle
Aorta situated anterior and to the right of the pulmonary artery
Ventricular septal defect
Auricular septal defect
Patent ductus arteriosus

Course of the Circulation

Systemic venous blood reaches the right heart and without oxygenation returns to the systemic circulation through the transposed aorta

Oxygenated blood reaches the left heart through the pulmonary veins and returns to the lungs from the left heart through the transposed pulmonary artery

Thus there are two entirely separate circulations This condition is incompatible with extra uterine life unless there *are associated abnormalities that permit shunts between* the two circulations

Associated lesions are ventricular septal defects auricular septal defects, and patent ductus arteriosus Any one or all of these associated lesions may be present These compensating shunts are necessary in order to maintain blood volume equilibrium

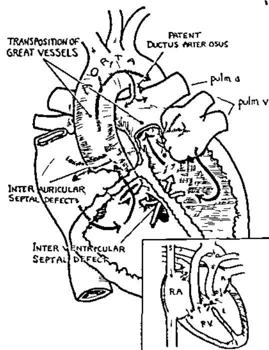


PLATE 17

Complete transposition of the great vessels associated *with ventricular and* auricular septal defects and a patent ductus arteriosus

Clinical Features

Cyanosis from birth
Progressive cardiac enlargement begins in early infancy
Poor growth and development
Enlarged liver
Basal systolic murmur

Electrocardiogram

Right axis deviation
P waves may be abnormal

Radiological Studies

Cardiac enlargement
Shadow cast by the great vessels narrow in A P view
Shadow cast by the great vessels broad in L A O view
There is usually evidence of pulmonary congestion
Angiocardiography demonstrates the transposed aorta and the vessels' anterior position

Simultaneous filling of aorta and pulmonary artery will be seen in the angiocardiogram if a ventricular septal defect is present

Laboratory

Arterial O_2 unsaturation
Hemoconcentration
Cardiac catheterization may be of value in determining the type and number of associated malformations

Prognosis

Poor—majority of patients with this malformation die in early infancy

Treatment

Experimental Creation of an extra cardiac shunt and an auricular septal defect in our experience, has been of little benefit



Fig 105



Fig 106



Fig 107

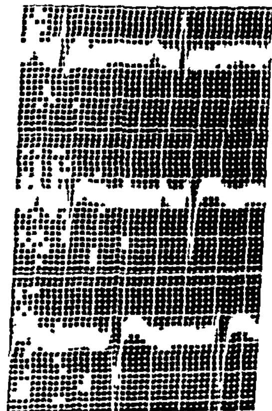


Fig 108

Complete Transposition of Arterial Trunks with "Riding" Tricuspid Valve Type IV of Spitzer

(Age 8 years)

Fig 105 Complete transposition of arterial trunks Notice the aorta in front and to the right of the pulmonary artery

Fig 106 Same case Posterior view of the heart showing the right atrium and the tricuspid valve Notice how the tricuspid valve extends over a defect in the ventricular septum and opens into both ventricles

Fig 107 A P roentgenogram of the chest Notice the left ventricular hypertrophy the broad supracardiac shadow and the increased pulmonary vascularity

Fig 108 E K G showing a pronounced left axis deviation The Q R S complex is upright in lead I and inverted in leads II and III

Fig 109 Complete transposition of the arterial trunks Sections of aorta and pulmonary artery Notice the thick pulmonary artery with a vegetative overgrowth on the intimal surface Elastic tissue stain (X 15)

Fig 110 Same case Section of lung showing two arteries with an almost complete occlusion of the lumen by concentrical layers of fibrous connective tissue Elastic tissue stain (X 200)

Fig 111 Same case Section of lung showing large and thick walled pulmonary arteries Elastic tissue stain (X 15)

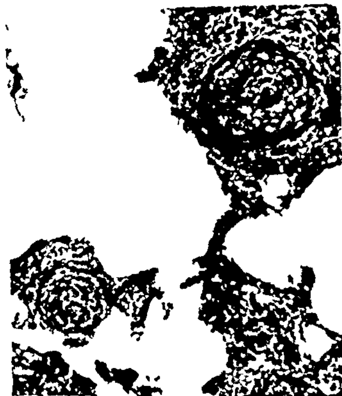


Fig 110



Fig 109



Fig 111



Fig 112

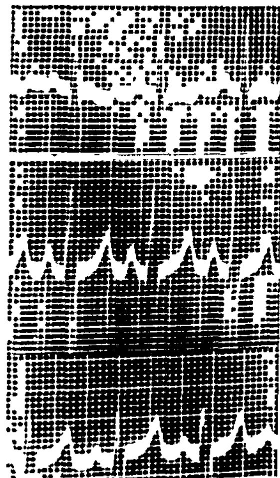


Fig 113



Fig 114



Fig 115

Complete Transposition of Arterial Trunks with Infundibular (Subaortic) Stenosis

(Age 3½ years)

Fig 112 Heart showing the aorta in front and to the right of the pulmonary artery Notice the infundibular (subaortic) stenosis present

Fig 113 E K G showing moderate left axis deviation The QRS complex is upright in lead I and diphasic in leads II and III Peaked P waves are present in lead II

Fig 114 Same case A P roentgenogram of chest Notice the cardiac enlargement and the vascular lung fields

Fig 115 Section of aorta and pulmonary artery Notice the pronounced thickness of the pulmonary artery (larger section) Elastic tissue stain (X 15)

Fig 116 Section of lung revealing very prominent and thick pulmonary arteries Elastic tissue stain (X 15)

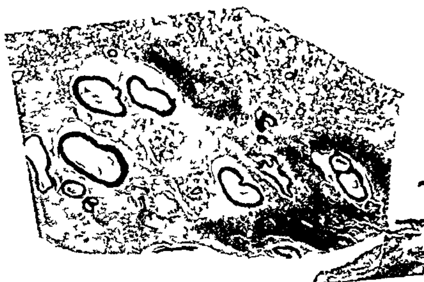


Fig 116



Fig 117



Fig 118



Fig 119

Complete Transposition of Arterial Trunks

(Age 6 weeks)

Fig 117 Heart and lungs Notice the aorta to the right and slightly anterior to the pulmonary artery

Fig 118 A P roentgenogram of the chest Notice the cardiac enlargement and the narrow supracardiac shadow

Fig 119 Angiocardiograph Notice the simultaneous visualization of the aorta and the pulmonary artery as a result of a ventricular septal defect The aorta originates in the right ventricle

Fig 120 Section of aorta and pulmonary artery Notice the very thick pulmonary artery (longer section) Elastic tissue stain (× 15)

Fig 121 Section of lung showing very large and thick pulmonary arteries Elastic tissue stain (× 15)



Fig 120



Fig 121



Fig 117



Fig 118



Fig 119

Ventricular Inversion

Pathology

Anatomical left ventricle and mitral valve are located beneath the normal right atrium
Anatomical right ventricle and tricuspid valve are located beneath the normal left atrium

Course of the Circulation

Normal

Pulmonary Valve Stenosis with Ventricular Inversion

(Age 1½ years)

Fig 124 Left side of the heart showing the left atrium the tricuspid valve and the anatomical right ventricle The aortic valve is situated behind the tricuspid valve The picture also shows the stenotic pulmonary valve with a vegetative process in it

Fig 125 A P roentgenogram of the chest Notice the large pulmonary artery and the avascular lung fields

Fig 126 Right side of the heart showing the right atrium the mitral valve and the anatomical left ventricle with an added infundibular portion

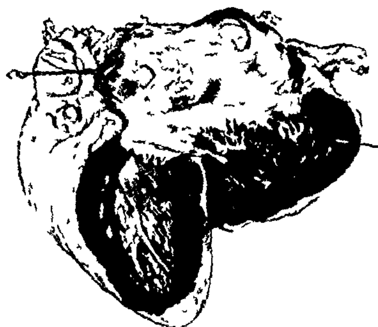


Fig 124



Fig 125



Fig 126

Corrected Transposition

Pathology

Aorta and aortic valve anterior
Pulmonary artery and pulmonary valve posterior
Each vessel arises from the corresponding ventricle

Course of the Circulation

Normal

Fig 122

Corrected Transposition

(Age 40 years)

Fig 122 *Heart from a 40 year old male* with corrected transposition of the arterial trunks The aorta is anterior to the pulmonary artery but originates from the left sided ventricle Notice the abnormal ap*pearance of the papillary muscles* Notice also the crista supraventricularis running from the base of the aortic valve to the anterior wall of the ventricle

Fig 123 Section of aorta and pulmonary artery showing normal *thickness* and relationship

Fig 123

6 Anomalies of the Aortic Arch

Coarctation of the Aorta

Pathology

Constriction of the aorta at or near the point of entry of the ductus arteriosus
- Infantile type—preductal
- Adult type—postductal

Preductal type usually associated with intracardiac anomalies

Course of the Circulation

Preductal type—right to left shunt through the ductus arteriosus

Postductal type—results in development of large collateral channels (internal mammaries, intercostals, anastomosis about the clavicle)

Clinical Features

A common cause of heart failure in acyanotic infant
Many cases asymptomatic, diagnosed on routine examination
Normal growth and development
Hypertension in upper extremities
Weak or absent femoral pulsations
Heart of normal size
Soft systolic basal murmur
Loud systolic murmur over the left chest posterior

Electrocardiogram

Left axis deviation
Left ventricular preponderance

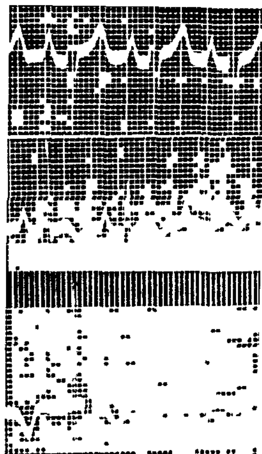


Fig 127

Fig 127 E K G showing peaked P waves in leads I and II and inverted P wave in lead III There are also marked right axis deviation and inverted T waves in lead II and III

Fig 128 Section of aorta and pulmonary artery Notice the thin pulmonary wall which reveals a vegetative process at one end

Fig 129 Section of lung showing very small and thin walled pulmonary arteries for a 14 year old Elastic tissue stain (× 15)



Fig 128

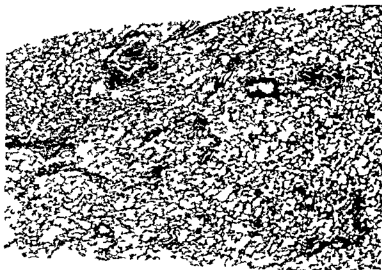


Fig 129

Coarctation of the Aorta

(Age 2½ months)

Fig 130 A P roentgenogram of the chest (pre-operative) from a case of coarctation of the aorta Notice the marked cardiac enlargement

Fig 131 A P roentgenogram of same case less than twenty four hours postoperative Notice the marked decrease in cardiac size

Fig 132 *A line drawing to represent the* coarctation and surgical repair in this case



Fig 131

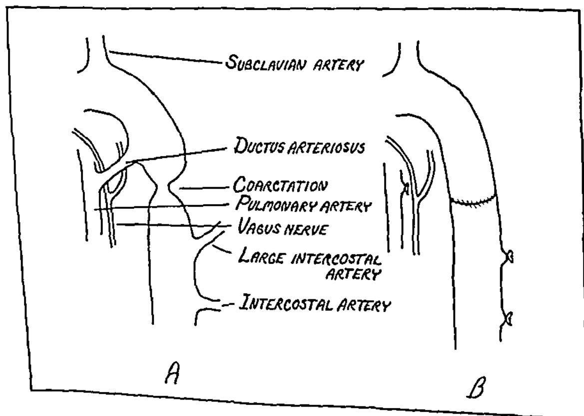


Fig 132

Radiological Studies

Heart of normal size
Notching of inferior rib margin (adults)
Arteriogram reveals coarcted area and the collateral vessels

Laboratory

Direct arterial pressure tracings from the brachial and femoral arteries confirm the hypertension in the upper extremities

Prognosis

Good

Treatment

Surgical—resection of constricted area with end to end anastomosis, the use of human arterial graft



Fig 130

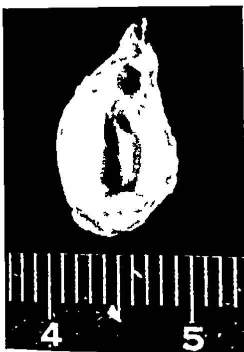
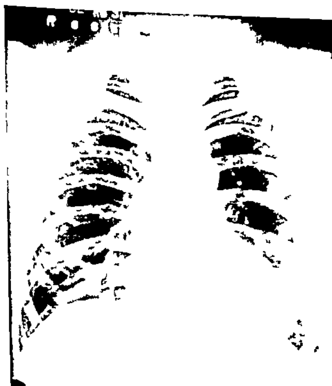


Fig 135



Fig 136



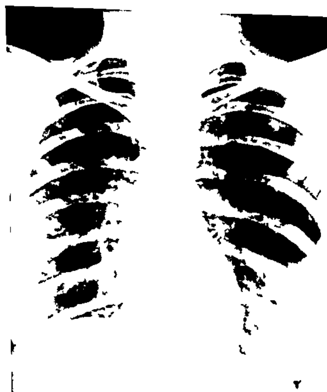


Fig 137

Fig 133

Coarctation of the Aorta

(*Age 22 years*)

Fig 133 Arteriogram from a case of coarctation of the aorta Notice the constricted area in the thoracic aorta and tortuous internal mammary artery

Fig 134 A line drawing to illustrate the location of the coarctation in this case and the type of surgical removal

Fig 135 Specimen of the same case Notice the marked constriction in the lumen of the aorta

Fig 136 Specimen of coarctation Notice the marked dilatation of the aorta distal to the coarctated area

Figs 137 and 138 A P roentgenograms from two cases of coarctation Notice notching of the undersurface of the ribs as seen in adults

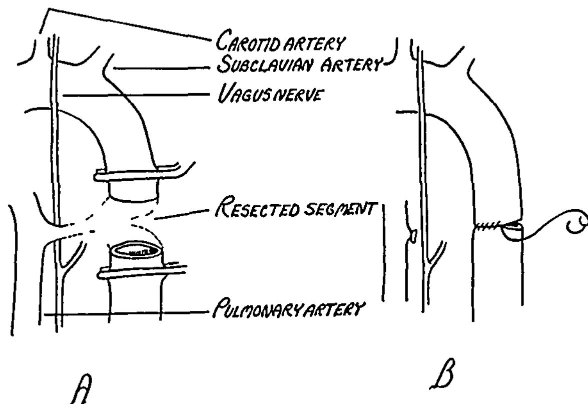


Fig 134



Fig 142

Pre Ductal Coarctation

(*Age 1 year*)

Fig 142 Heart and great vessels from a one year old child with infantile coarctation Notice the diffuse narrowing of the arch of the aorta from the point of origin of the left carotid to the point of entrance of the ductus arteriosus



Fig 139

Coarctation of the Aorta with rupture of the wall below the point of constriction

(11 year old male)

Fig 139 Aorta from an 11 year old male with coarctation Notice the sharply localized constriction the large upper intercostal arteries and the insertion of the ligamentum arteriosum

Fig 140 A P roentgenogram from the same case The heart is normal in size and configuration Notching of the ribs is not present

Fig 141 Specimen from same case that shows rupture of the aorta which occurred below the area of coarctation and into esophagus



Fig 140



Fig 141

Arterial Rings

(*Illustrations on page 88*)

Pathology

Constricting arterial ring around the trachea and esophagus
Arterial ring usually consists of a double arch or a right aortic arch with a left descending aorta The ductus or ligamentum arteriosus completes the constricting ring

Course of the Circulation

There is usually no associated intracardiac anomaly
The hemodynamics of the circulation are not altered

Clinical Features

Increased incidence of respiratory infections in early childhood
Dysphagia in early childhood
There may be respiratory distress
No cardiac murmurs

Electrocardiogram

Normal

Radiological Studies

Barium swallow usually reveals a right arch or evidence of a double arch A retro esophageal vessel is usually discerned

Prognosis

If the patient survives the first year of life the prognosis is usually excellent however stridor, dysphagia and repeated respiratory infections may make surgical intervention necessary

Treatment

Surgical

Patent Ductus Arteriosus

Pathology (Plate 18)

The ductus arteriosus has its origin at the bifurcation of the pulmonary artery, and its entrance into the aorta is two or three centimeters distal to the point of origin of the left subclavian artery

Course of the Circulation

The higher pressure in the aorta produces a left to right shunt (aorta to pulmonary) through the ductus

Clinical Features

More common in the female
Growth and development may be normal or retarded
Heart normal size
Continuous roaring murmur in 2nd left I C S outside the area of cardiac dullness
A marked thrill present in the same area
Peripheral signs of aortic insufficiency

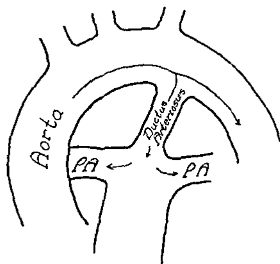


PLATE 18
Patent ductus arteriosus

Electrocardiogram

Usually normal

Radiological Studies

Heart usually normal size or slightly large
The left ventricle may be prominent
Increased vascularity of the lung fields
Hilar pulsations noted on fluoroscopy
Appearance of the pulmonary conus not consistent
In atypical cases arteriograms may be of value in diagnosis—dye can be seen entering the lung fields from the systemic circulation

Laboratory

Catheterization reveals an increase in the oxygen content of the pulmonary artery sample as compared to the right ventricular sample

Prognosis

Good, complications usually do not develop until adult life

Treatment

Surgical closure of the ductus

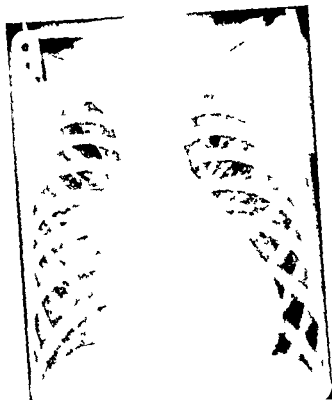


Fig 146

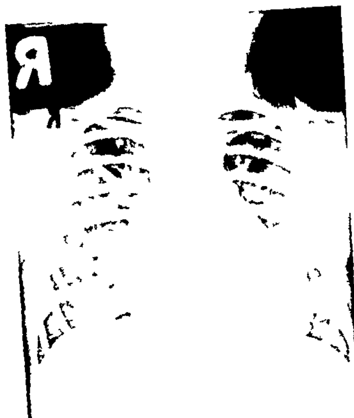


Fig 147

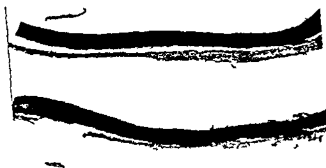


Fig 148



Fig 149



Fig 143

Patent Ductus Arteriosus

(5 year old male)

Fig 143 Heart with a patent ductus arteriosus The heart is enlarged bilaterally

Fig 144 The electrocardiogram (standard leads) from the same case reveals a normal tracing

Figs 145, 146 and 147 A P roentgenograms from children between the ages of seven and nine with a typical patent ductus arteriosus Notice the difference in the shadow cast by the pulmonary conus in the three cases Also notice that heavy lung markings are common to all three

Fig 148 Sections of aorta and pulmonary artery from a case of patent ductus arteriosus Notice the hypertrophy of the pulmonary arterial wall which is as thick as the aortic wall Elastic tissue stain (× 15)

Fig 149 Lung section from a case of patent ductus arteriosus showing the large and thick pulmonary arteries Elastic tissue stain (× 15)

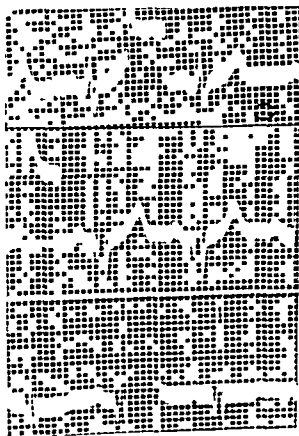


Fig 144

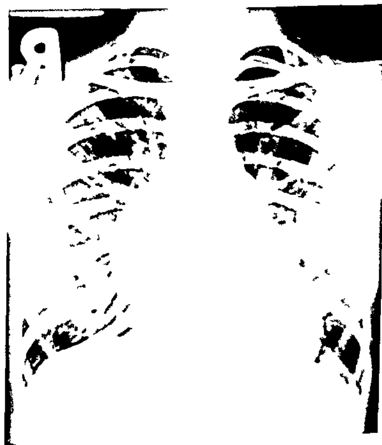


Fig 145

7 Ebstein's Malformation of the Tricuspid Valve

Pathology

This condition consists of a malformation of the tricuspid valve in which there is marked fusion of the leaflets to each other and to the ventricular wall and also a downward displacement of the posterior and lateral aspects of the valve Moreover, the proximal portion of the right ventricle is incorporated into the huge right auricle and it communicates with the distal (outflow) portion of the ventricle through a large opening in the anterior leaflet of the tricuspid valve In most cases there is an associated auricular septal defect

Course of the Circulation

The malformation of the tricuspid valve and the right side of the heart results in an increased pressure and a marked dilatation of the *right atrium* Cases with an auricular septal defect develop a marked right to left shunt, with decrease in the pulmonary blood flow and associated cyanosis

Clinical History

Retardation of growth and development

Delayed onset of cyanosis disproportion between degrees of cyanosis and dyspnea Limitation of activity with easy fatigability No history of squatting Frequent episodes of paroxysmal tachycardia

Physical Examination

Mild to severe cyanosis Cardiac enlargement, mostly right atrium and ventricle Distant heart sounds systolic murmur, gallop rhythm



Fig 150



Fig 151

Arterial Ring Around Trachea and Esophagus

Fig 150 A P roentgenogram of chest from a case of vascular ring age 6 months Notice that the barium swallow reveals a bilateral compression of the esophagus

Fig 151 L A O roentgenogram of the chest from the same case Notice the anterior displacement of the barium filled esophagus by the retroesophageal vessel

7 Ebstein's Malformation of the Tricuspid Valve

Pathology

This condition consists of a malformation of the tricuspid valve in which there is marked fusion of the leaflets to each other and to the ventricular wall and also a downward displacement of the posterior and lateral aspects of the valve Moreover the proximal portion of the right ventricle is incorporated into the huge right auricle and it communicates with the distal (outflow) portion of the ventricle through a large opening in the anterior leaflet of the tricuspid valve In most cases there is an associated auricular septal defect

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The malformation of the tricuspid valve and the right side of the heart results in an increased pressure and a marked dilatation of the right atrium Cases with an auricular septal defect develop a marked right to left shunt with decrease in the pulmonary blood flow and associated cyanosis

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Physical Examination

Mild to severe cyanosis Cardiac enlargement mostly right atrium and ventricle Distant heart sounds systolic murmur gallop rhythm

Electrocardiogram

Prolonged auricular ventricular conduction Right bundle branch block High and peaked P waves At times, paroxysmal supraventricular tachycardia

Radiological Studies

Clear lung fields Concave pulmonary conus Cardiac enlargement, especially right atrium and right ventricle *Angiocardiography reveals pooling of the dye in the enlarged* right auricle for too long period of time

Laboratory Studies

Prolonged circulation time Decreased pulmonary blood flow, with right to left flow through auricles Hemoconcentration Arterial oxygen unsaturation

Treatment

Medical No surgical treatment at present

Prognosis

Poor



Fig 152



Fig 153

Ebstein's Malformation of the Tricuspid Valve

(Age 29 years)

Fig 152 Heart and lungs in a case of Ebstein's malformation Notice the marked dilatation of the right side of the heart and the small size of the pulmonary artery

Fig 153 Ebstein's malformation Anterior view of the right ventricle showing a marked dilatation of the pulmonary conus and a large opening at the base of the anterior leaflet of the tricuspid The pulmonary valve can also be seen

Fig 154 Ebstein's malformation Posterior view of the right ventricle and atrium showing the enormous dilatation of the former Notice the marked displacement of the posterior leaflet of the tricuspid valve toward the apex of the heart Notice also the incorporation to the large right atrium of the proximal portion of the right ventricle In the center of the picture there is the large defect through the anterior leaflet of the tricuspid On the upper left the patent foramen ovale can be seen

Fig 155 Ebstein's malformation E K G showing paroxysmal supraventricular tachycardia right ventricular preponderance and right bundle branch block



Fig 154

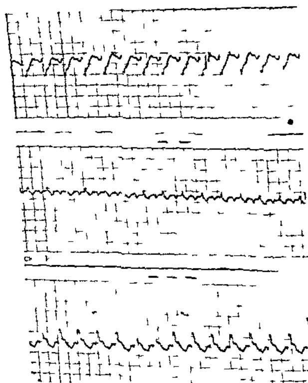


Fig 155



Fig 156

Fig 156 Ebstein's malformation A P radiograph of the chest showing a marked increase in cardiac size and normal pulmonary vascular markings The pulmonary conus is not prominent

Fig 157 Ebstein's malformation L A O (almost lateral) radiograph of the chest Notice the very large right ventricle which reaches the anterior chest wall



Fig 157

